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Mesenchymal Chondrosarcoma of the Foot, an Unusual Location: Case Report and Review of the Literature

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We present a case of primary mesenchymal chondrosarcoma of the proximal phalanx of the first toe. The bones of the foot represent an infrequent primary site for this neoplasm. The tumour consisted of layers of undifferentiated round cells with scanty cytoplasm and hyperchromatic nuclei. The presence of brain, lung, and left auricle metastasis was demonstrated, and the patient died due to brain edema 18 days after admission. Mesenchymal chondrosarcoma is a rare tumor that more frequently involves the pelvic bones, the femur, and the humerus. To our knowledge, only nine cases of primary mesenchymal chondrosarcoma arising from the bones of the foot have been previously reported, with none involving the phalanx of the toe.

Key words: mesenchymal chondrosarcoma, bone tumors, foot tumors, auricle metastasis

INTRODUCTION

Chondrosarcomas represent only 10% of all malignant bone tumors [1]. They most often appear in males between 40 and 70 years of age and usually arise from the proximal end of the long bones. The most common signs and symptoms are the presence of a mass and pain [1].

There are four morphological types of centrally located chondrosarcomas: classical, dedifferentiated, clear-cell, and mesenchymal chondrosarcomas [2]. This last group constitutes ~13% of the chondrosarcomas, affects patients in the second and third decades of life, and frequently involves the mandible, vertebra, pelvic bones, and femur, with a poor prognosis [3].

As far as we know, there have been very few reports of primary mesenchymal chondrosarcoma originating in the bones of the foot [3–7], with the os calcis being the most frequent location described. Here we report the first case, to our knowledge, sited in a phalanx and review the literature.

CASE REPORT

A 22-year-old man was admitted to our hospital in August 1992 because of a 9-month history of progressive dyspnea and cough, with holocraneal cephaledema that had increased over the previous 2 months. Medical history revealed that he had suffered a fracture in the first toe of the right foot 2 years before, which had not received medical attention and sometimes caused local pain.

On physical examination, he was in good general health and had pronounced shortening of the first toe of the right foot with a hard swelling of the first metatarsophalangeal joint. Other examination findings were an absence of breath sounds in the lower half of the left lung and a slight central paresis of the right facial nerve. The remainder of the examination was completely normal.

The only laboratory finding was a lactic dehydrogenase of 540 U/1. HIV serology was negative. Right foot roentgenogram showed an osteolytic lesion with destruction of the proximal phalanx of the first toe (Fig. 1). Chest roentgenograms demonstrated a 12 cm diameter round, well-defined mass in the lower lobe of the left lung. Thoracic computerized tomography was performed confirming the presence of this mass, which contained a great number of small and confluent calcifications in its centre, involved the mediastinum, and invaded the left auricle (Fig. 2). Two small nodes, ~1 cm in size, were also seen in the upper lobe of the left lung. Verification of a 5 cm mass occupying the 75% of the left auricle cavity

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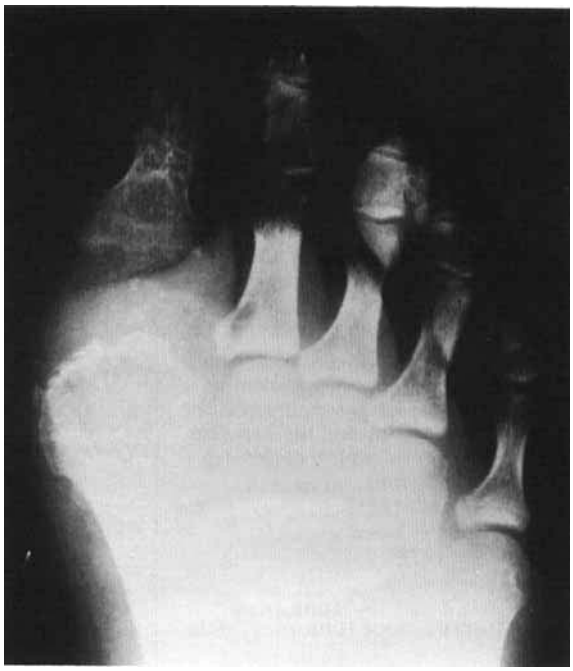

Fig. 1. Right foot roentgenogram revealing an osteolitic lesion, with destruction of the proximal phalanx of the first toe.

was obtained by an echocardiogram. Bronchoscopy found extrinsic compression at the entrance of the lower lobe bronchus of the left lung.

A transbronchial biopsy as well as a biopsy of the osteolytic lesion of the proximal phalanx of the toe were performed. On both microscopic examinations, the tumor consisted of sheets of undifferentiated round cells without any specific arrangement. These cells had scarce cytoplasm and oval, hyperchromatic nuclei. A sudden transition from undifferentiated mesenchymal cells to islands of mature cartilage was observed (Fig. 3). A diagnosis of mesenchymal chondrosarcoma was made.

A few days after admission, the condition of the patient deteriorated presenting increased cephaledema and drowsiness. The fundoscopic examination showed a bilateral papilledema and computerized tomography demonstrated two nodular lesions in the right cerebral hemisphere, surrounded by signs of cerebral edema. Antiedema therapy was started without improvement, and the patient died 18 days after admission.

DISCUSSION

We have described a case of mesenchymal chondrosarcoma located in the proximal phalanx of the first toe, which had been unnoticed until the patient's admission and in which disseminated metastasis were prominent presenting features and caused the death of the patient a few days later.

Mesenchymal chondrosarcoma is a rare tumour arising both in bone and soft tissues. Similar to conventional chondrosarcoma, it has a slight predominance in males, but although the average age of patients with ordinary chondrosarcoma is 40 years, those with the mesenchymal type have an age of ~25 years [6,8,9].

Clinical features of mesenchymal chondrosarcoma are not different from other bone tumours. Pain, swelling, and a mass are the most common findings [5]. The skeletal sites of involvement are frequently the pelvic bones, the femur, and the humerus. In extraskeletal sites, the cranial or spinal meninges are the most frequently affected [3]. To our knowledge only nine cases of primary mesenchymal chondrosarcoma involving the bones of the foot have been previously reported [3,5–7,10]. Most arose from the bones of the tarsus, and none has been reported originating in the phalanx of a toe.

The roentgenograms usually show irregular, lytic lesions with granular, mottled calcifications and invasion of the soft tissues. Periosteal reaction is less common than in other bone tumours such as osteosarcoma [7,8]. Similar findings can be noted in computerized tomography, which usually provides an accurate evaluation of tumour borders as well as signs of soft tissue involvement by the sarcoma [1].

In typical lesions, the histologic features of mesenchymal chondrosarcoma (a combination of undifferentiated round cells and chondroid component) are very characteristic. However, diagnosis may at times be difficult. The histologic differential diagnosis includes malignant haemangiopericytoma, Ewing's sarcoma, small cell osteosarcoma, and lymphoma. A search for areas containing chondroid matrix may help to solve the problem.

As shown in the case reported, mesenchymal chondrosarcoma has a highly aggressive behaviour. Radical surgery is accepted as the main method of treatment but has many limitations since many tumors cannot be removed adequately owing to distant metastasis or local extension to unresectable sites [11]. Radiation therapy and chemotherapy may be used as an adjunctive treatment after initial surgery or may be suitable for lesions that are not candidates for ablative surgical treatment, although these therapies have not demonstrated significant improvement in the results [3].

Prognosis is markedly worse for mesenchymal chondrosarcoma than for the classic type. Survival analysis revealed a 46% 2-year and a 35% 5-year survival rate for the first [6], and a 67% 5-year survival rate for the second [12]. More than half of the patients with mesenchymal chondrosarcoma develope distant metastasis with the lung being the most frequent site [3], as occurred in the case reported.

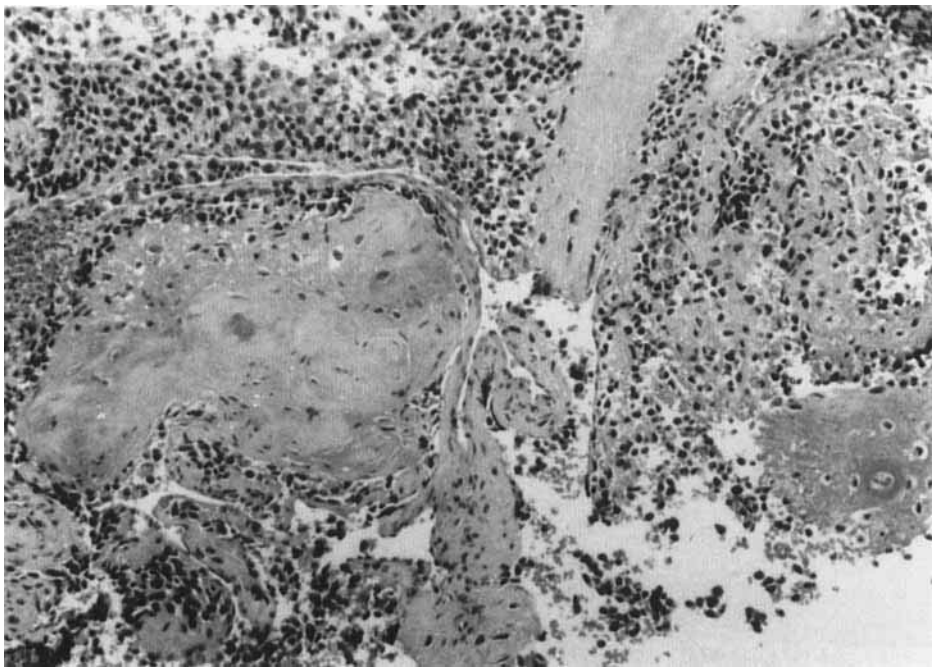

Fig. 2. Thoracic computerized tomography demonstrates a 12 cm, well-defined mass in the lower lobe of the left lung, with confluent calcifications in its centre and invasion of the left auricle cavity.

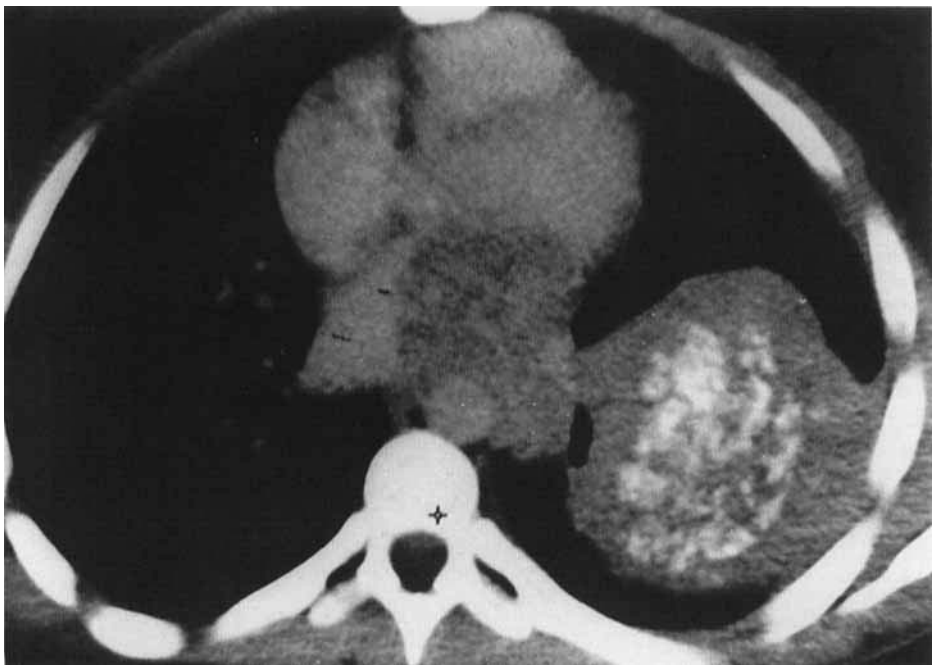

Fig. 3. Proximal phalanx of the first toe biopsy shows undifferentiated mesenchymal cells with sudden transition to mature cartilage (Hematoxylin-eosin × 40).

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